Carrier Screening for Sickle Cell Disease

Slides Previously Presented to ACHDNC by
Lanetta B. Jordan, M.D., M.P.H., M.S.P.H.
Chief Medical Officer
Sickle Cell Disease Association of America, Inc.

R. Rodney Howell, M.D.
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Screening, Follow-up, Health Education

- 2006 Universal Hemoglobinopathy Screening of Newborns (90% of all newborns screened since 1993)

Screening for Disease

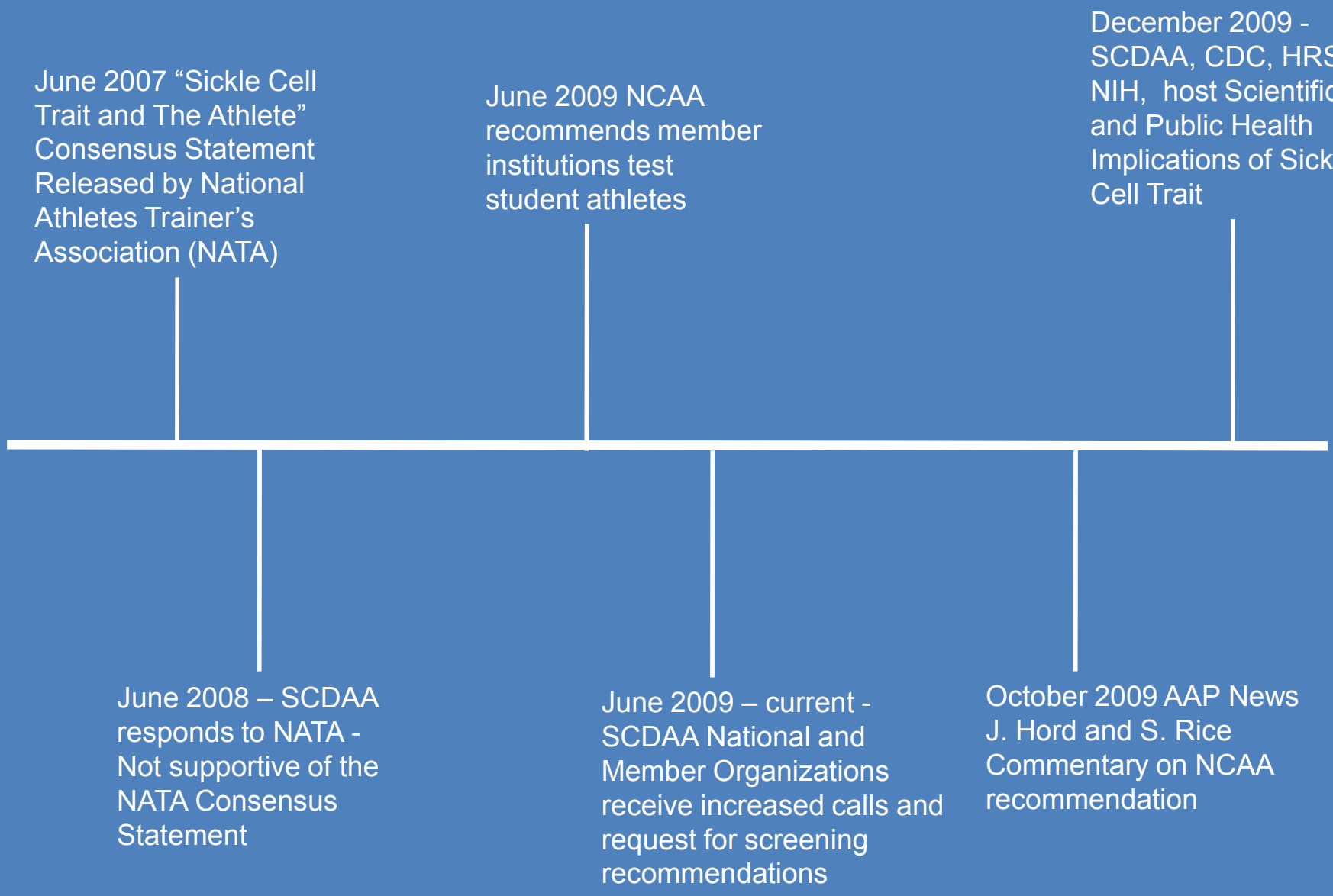
Screening to identify confirmed cases to initiate medical care, vaccination against S. pneumoniae, H. influenza type b, Meningococcus type c infections, educate parents on health maintenance and health risks;

Carrier Screening - screening in asymptomatic individuals for genetic pre-disposition for disease/condition (no longer benign)

- Carrier Status

State variability in carrier status recording of test results and parental notification;

- Lack of agreed upon clinical evidence defining health risks associated with carrier status, cost /benefit challenge?


June 2007 “Sickle Cell Trait and The Athlete” Consensus Statement Released by National Athletes Trainer’s Association (NATA)
June 2008 – SCDAA responds to NATA - Not supportive of the NATA Consensus Statement
June 2009 NCAA recommends member institutions test student athletes
June 2009 – current - SCDAA National and Member Organizations receive increased calls and request for screening recommendations
October 2009 AAP News J. Hord and S. Rice Commentary on NCAA recommendation
December 2009 - SCDAA, CDC, HRSA, NIH, host Scientific and Public Health Implications of Sickle Cell Trait

State of the Evidence for Health Outcomes Associated with Sickle Cell Trait

Assessment of Significant Relative Risk

Retrospective Analysis (1977 – 1981)

Non-traumatic deaths
2 million military recruits

AA Recruits with HbS (N=13 deaths)	RR 30
AA and other Recruits without HbS (N=5 deaths)	RR 3

AA = African American
RR = Relative Risk

Reference – J. Kark

State of the Evidence for Health Outcomes Associated with Sickle Cell Trait

Assessment of Significant Relative Risk?

Intervention Trial (1982 – 1991)
Endpoint = Prevent Exercise Related Death

1.8 million basic training recruits

Intervention = Strict protocol to prevent exercise health illness/injury

Outcome = Not one of the 13 predicted deaths occurred

Reference – J. Kark

State of the Evidence for Health Outcomes Associated with Sickle Cell Trait

Assessment of Significant Relative Risk

Intervention Trial (1982 – 1991)

Conclusions

1) Prevention of exercise related death did not require identification of sickle cell trait, as prevention, diagnosis, and treatment of exercise heat related illness/injury are unrelated to hemoglobin type;

2) Exertional heat illness is a preventable factor contributing to sudden exercise related death in persons with sickle cell trait.

State of the Evidence for Health Outcomes Associated with Sickle Cell Trait

Assessment of Significant Relative Risk

Evolving Military Policy (1960 – current)

Conclusions

1) Evidence supports sickle cell trait as an increased risk for exertional health illness or injury, likely with contribution from still unidentified genetic polymorphisms;

2) Sickle cell trait does not exclude military personnel from duty in the Army; Air Force, Navy and Marines screen for certain military occupations;

3) Preventive measures can reduce exertional health illness or injury.

Carrier/Trait Re-Screening, Follow-up, Health Education

COST

- 400,000 collegiate athletes
- 8 million high school athletes

Sickledex test is inappropriate screening test
Hemoglobinopathy electrophoresis

College	$20,000,000
High school	$400,000,000

Such costs will likely result in re-screening of targeted groups

Reference – Hord and Rice 2009